

Prolapsed mitral cusps in atrial septal defect *An erroneous radiological interpretation*

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Forty patients with simple atrial septal defect had left ventriculograms using cineangiography and/or Elema or Sircam still films.

All had open heart surgery and careful scrutiny of the mitral valve by experienced surgeons. Prolapse of the posterior cusp was diagnosed from angiography in 28 patients but was confirmed at operation in only 5. Another 5 had a different mitral valve abnormality and the remaining 18 had no clinical signs to suggest mitral valve dysfunction after operation. It is concluded that though true ballooning or prolapse of the mitral cusps may coexist with simple atrial septal defect, its presence is overdiagnosed from the appearances of left ventriculography on anteroposterior, lateral, and right anterior oblique views. Left ventriculography was equally unreliable in the exact recognition of other anatomical abnormalities of the mitral valve in secundum defect, but accurate when the mitral valve was pronounced to be structurally normal.

The association of ballooning of the cusps of the mitral valve with atrial septal defect has been documented from angiographic studies (McDonald *et al.*, 1971; Pocock and Barlow, 1971; Epstein and Coulshed, 1973; McKay and Yacoub, 1973; Schwartz *et al.*, 1975). The patient with an atrial septal defect who has left ventriculography is often the one in whom a mitral valve abnormality is suspected, either because of suggestive clinical signs or because of unexplained left axis deviation on the electrocardiogram. Thus, there has been the temptation to assume that the unusual appearance of the mitral valve seen on the angiocardigram represents a pathological abnormality.

In order to test the accuracy of left ventriculography in diagnosing prolapsed mitral cusps in simple atrial septal defect, the angiocardigraphic images have been correlated with the anatomy of the mitral valve carefully examined at open operation by experienced surgeons.

Subjects

Forty patients with simple septal defect had left ventriculography before open heart surgery. Patients were aged 4 to 64 years at the time of

operation: 13 were less than 10 years, 12 were between 11 and 30, and 15 were over 30 years.

Each patient was operated on using cardiopulmonary bypass and the surgeon specifically examined the mitral valve by palpation when the left ventricle was beating, and afterwards by inspection when the aorta was clamped. When the defect was a small one, the atrial septum was opened more widely to permit complete examination of the mitral cusps and subvalvar mechanism when the left ventricle was perfused.

The atrial septal defect was in the fossa ovalis in 33, in the posterior septum opposite to the inferior vena caval opening in 5, and in the sinus venosus in 2. It was closed by direct suture in 28, by a pericardial patch in 11, and by a 'dacron' patch in one.

Eighteen patients were investigated because of physical signs which might have been caused by a mitral valve abnormality: an apical systolic murmur in 16, a variable praecordial 'honk' in one, and a late systolic click in the other; in another there was suspected aortic valve disease. A further 5 had isolated left axis deviation on the electrocardiogram but no mitral murmur, and 16 patients with 'uncomplicated' secundum atrial septal defects were included as 'controls'.

Method

In 31, the left ventricle was entered from the left atrium, the catheter having been passed through

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Received for publication 15 April 1977

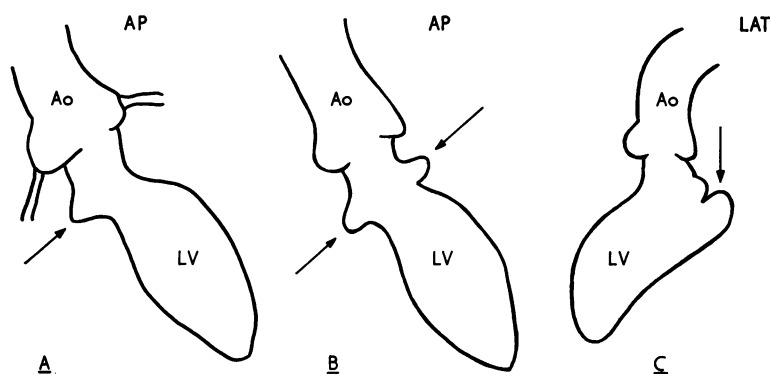


Fig. 1 Diagrams of left ventriculograms in systole (A) and (B) anteroposterior view, and (C) lateral view. Arrows show bulges interpreted as prolapsed mitral cusps.

the atrial septal defect. In 9, the left ventricle was catheterised retrogradely from a brachial or femoral artery. Direct anteroposterior and lateral views were taken in patients using Elema or Sircam films in 27 and cineangiography in 6; 17 had cineangiograms in the right anterior oblique position (RAO) (Honey *et al.*, 1969), and 10 had both investigations. The angiograms were examined by the authors and radiologists separately, and the anatomy of the mitral valve assessed as in previous studies (Criley *et al.*, 1966; Grossman *et al.*, 1968). Anteroposterior and lateral views were chosen in this study as they are the best for distinguishing atrioventricular defects from simpler forms of atrial septal defect (Somerville and Jefferson, 1968); in the first 10 patients this was the main clinical differential diagnosis. The same technique was used electively in the other patients in order to examine the diagnostic accuracy of the investigation.

In the anteroposterior view of the biplane angiocardio-gram, prolapsed (or 'ballooned') posterior cusp of the mitral valve was diagnosed when there was a convexity of the radio-opaque structure appearing on the right border of the left ventricle at the junction of the upper and middle thirds, or on both borders (Fig. 1A and 1B). In the lateral view, this appeared as a bulge in the lower (caudal) part of the mitral valve (Fig. 1C).

Prolapsed anterior cusp of the mitral valve was diagnosed when a radio-opaque structure beneath, and in continuity with, the aortic valve bulged excessively in a posterior direction in systole in the lateral view; this could not be identified in the anteroposterior view. In the right anterior oblique view, a bulge like a 'doughnut' on both sides of the aortic valve also suggested anterior cusp prolapse (Fig. 2). However, there is frequently overlapping of anterior and posterior cusps in this projection, so that it is uncertain if this appearance should be

interpreted as a radiological sign of anterior cusp prolapse without other views, which are necessary to establish which of the mitral cusps is prolapsing.

Other radiographic abnormalities of the mitral valve were noted when seen. Grading of severity of mitral regurgitation was not attempted as the catheter was passed through the mitral valve in many of the patients, and as the purpose of this study was to correlate the angiographic assessment of the pathological anatomy of the mitral valve with the surgical findings.

Results

The results of the study are summarised in Fig. 3.

In 9 patients, both cusps of the mitral valve were considered to be normal from the appearances of the left ventriculograms. In these cases no 'bulges' were seen on the medial or lateral borders of the left ventricle and the surgeon found a normal mitral valve in all of them. Left ventriculography

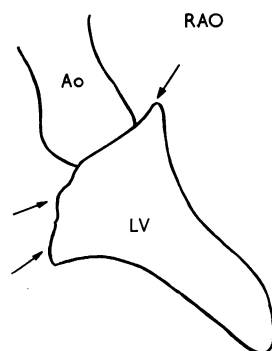


Fig. 2 Diagram of left ventriculogram (cine) in the right anterior oblique view, showing 'doughnut' appearance and suggesting prolapse of mitral valve cusps.

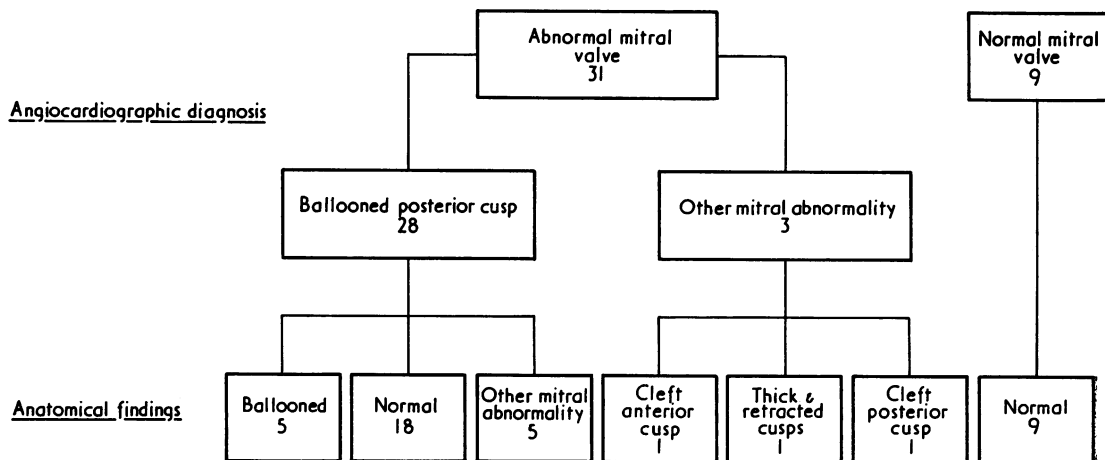


Fig. 3 Angiocardiographic assessment of anatomical state of mitral valve as seen from left ventriculography in 40 patients, related to findings at operation.

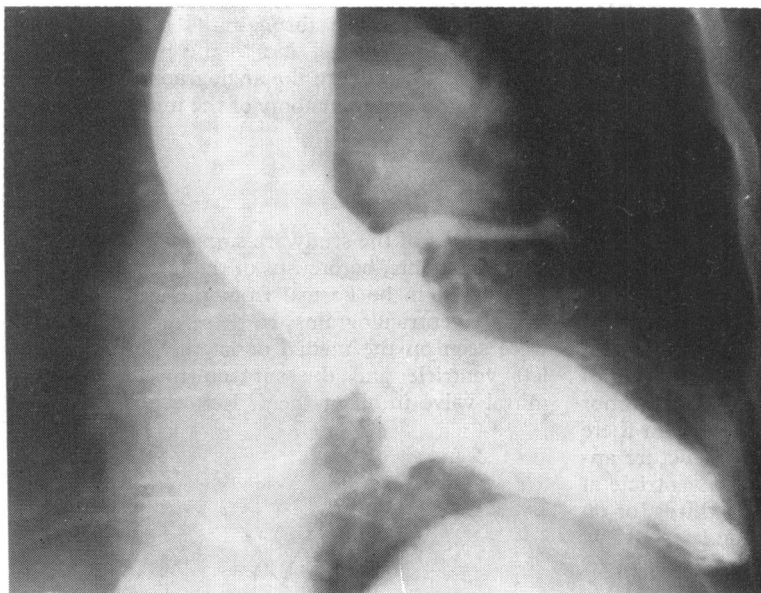


Fig. 4 Anteroposterior view left ventriculogram in a 56-year-old woman with left axis deviation, interpreted as showing prolapsed posterior cusp. The mitral valve was normal to inspection and testing at operation. Echocardiogram 3 years after operation did not show prolapsed cusps.

had been performed because of an apical systolic murmur in 4, left axis deviation in 2, and in 3 'controls'. None had signs suggesting mitral abnormalities after closure of the defect; all had a systolic ejection sound in the pulmonary area, thought to originate in the dilated pulmonary artery. These 9 patients were all under 14 years of age.

In all the other 31 cases, the left ventriculogram was interpreted as showing an abnormal mitral valve by conventional diagnostic criteria. Prolapse of the posterior leaflet was diagnosed in 28 (Fig. 4); 2 of these also had the appearance of redundant ballooned anterior cusp. In 18 out of the 28, the mitral valve was found to be normal at operation (Fig. 4 and 5) and there were no signs of mitral

dysfunction after closure of the atrial septal defect.

Only 5 of the 28 patients, aged 13, 36, 45, 50, and 56 years, at operation, had evidence of true prolapse of the posterior cusp, which was associated with redundancy of the anterior cusp in 2. The youngest patient, aged 13, had a variable praecordial 'honk' and ventricular ectopics (Fig. 6), and 2 of the 4 older patients had definite apical late systolic murmurs.

A further 5 patients in whom the left ventriculogram was thought to show a prolapsed posterior mitral cusp were found at operation to have different pathological abnormalities of the mitral valve. One patient had a cleft anterior cusp, another had rheumatic mitral stenosis, a third had ring dilatation with central regurgitation (Fig. 7A and 7B), a fourth had calcium in the anterior leaflet, which had spread from the aortic valve (Fig. 8), and

the fifth had a large and redundant anterior cusp with a shortened and rolled posterior cusp.

In the 3 patients where the mitral valve was considered to be angiographically abnormal but not prolapsing, one had regurgitation through a cleft anterior cusp, another had a completely cleft posterior cusp, and the third had rheumatic mitral valve disease with thickening and retraction of both cusps.

Discussion

The reported incidence of angiographic prolapse of the mitral valve in simple atrial septal defect varies from 8 to 37 per cent (Betriu *et al.*, 1975). In the current series, angiographic assessment by experienced radiologists using the same diagnostic criteria as in other series, showed it to be present in 28 cases (70%), but its presence could only be confirmed at operation in 5 (12%). Clinical examination after operation showed no mitral signs in 18 where no abnormality was found, and obvious clinical signs of mitral dysfunction in those 5 where the diagnosis was confirmed at operation. Thus, it appears from these findings that there is a problem in the interpretation of the left ventriculogram in patients with simple atrial septal defect when using anteroposterior, lateral, or right anterior oblique

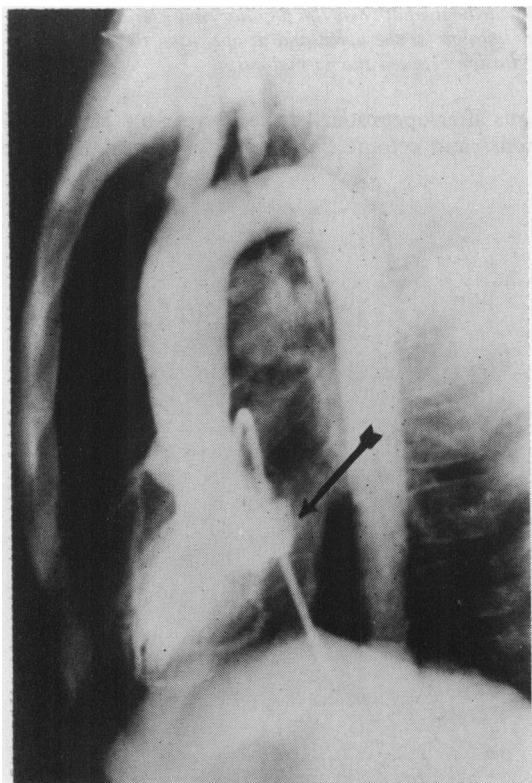


Fig. 5 Lateral view. Left ventriculogram from a 7-year-old boy. The anteroposterior view was thought to be normal but possible prolapse of the posterior cusp was questioned from the appearance of the cusps on the lateral view (arrow). The mitral valve was normal at operation.

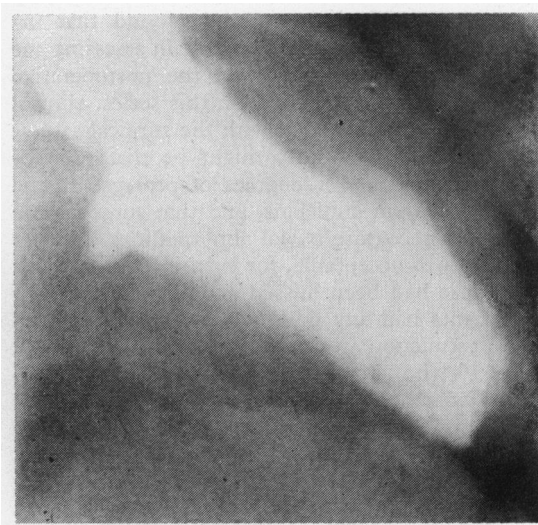


Fig. 6 Right anterior oblique left ventriculogram (cine), interpreted as showing ballooning of the posterior cusp of the mitral valve. At operation a redundant prolapsing posterior cusp was found. This patient was a 13-year-old girl with ventricular ectopic beats, right axis deviation, and a variable apical 'honk'.

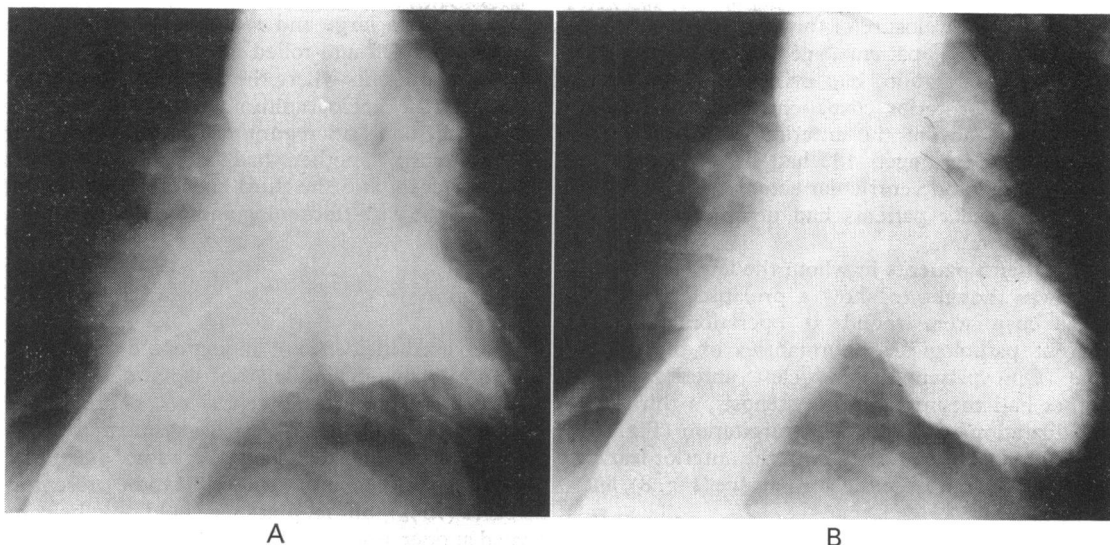


Fig. 7 Anteroposterior left ventriculograms in (A) systole, (B) diastole. [These were interpreted as showing prolapsed posterior cusp with mitral regurgitation and possible hypertrophic cardiomyopathy as the papillary muscles appeared to be enlarged. This patient was a woman aged 20 with late systolic murmur at the apex, and at operation the mitral ring was found to be dilated, allowing small central jets of regurgitation between the normal cusps.

views. Perhaps if the left anterior oblique view had been used, as suggested by Ranganathan *et al.* (1976), the findings would have been more reliable.

It could be argued that since no actual measurement of cusp size was made at operation, minor degrees of redundancy or prolapse were missed by the surgeons. We have certainly found that inexperienced surgeons are unreliable in assessing the mitral valve at operation, and the postoperative events have shown this; but, in this series, clinical findings correlated exactly with the surgical assessment. Another possibility might be that radiologists can detect lesser degrees of prolapse of the mitral valve than clinicians, and that surgeons are not able to recognise trivial abnormalities. Neither explanation is acceptable, for even if an early stage of prolapse had been missed at operation, none of the patients had any postoperative signs of mitral valve dysfunction in the early years of follow-up, and in 10 who later had echocardiograms, no prolapse was shown. These patients should not be labelled as having an abnormal mitral valve since this implies a need for lifetime protection against infective endocarditis, as well as presaging other possible developments in the natural history which may never occur. What will happen to the mitral valve in patients with a closed atrial septal defect 10 to 20 years later remains to be seen.

It is accepted that, occasionally, true prolapse of mitral cusps may occur with atrial septal defect, as in 5 patients in our series, all of whom had physical

signs after operation. In the 13-year-old girl with a 'honk' and ectopic beats, it is presumed that the

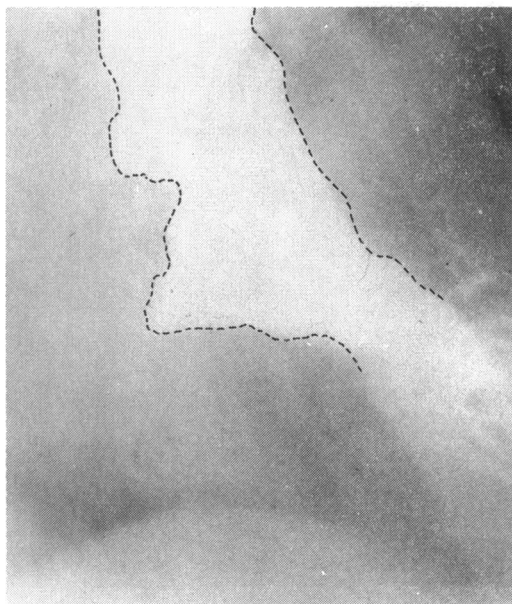


Fig. 8 Right anterior oblique left ventriculogram (cine) interpreted as showing prolapse of mitral valve. This patient was a 24-year-old man with right axis deviation who was found to have aortic valve stenosis and a normal mitral valve.

lesion was congenital. The other 4 with proven prolapsed mitral cusps were over 30 years old, thus raising the possibility that the abnormality might have been acquired. How this occurs must be speculative. It is possible that the anatomical characteristics of the mitral valve are altered in atrial septal defect as a consequence of disturbed left atrial blood flow patterns and the abnormal position of the mitral valve in a heart which is rotated in a clockwise direction as a result of right ventricular dilatation. Thickening and calcification of the mitral cusps are commonly found in elderly patients with atrial septal defect; this is not rheumatic or infective in origin but may be secondary to long-standing turbulent flow. Whatever causes these changes may also cause the apparent or actual prolapse of mitral cusps; this would account for the age at which these abnormalities truly coexist. The anatomy of the normal mitral valve is variable (Anton Becker, 1976, personal communication), and with a certain type of chordal insertion there could be a predisposition to prolapse of mitral leaflets subjected to unusual stresses. This predisposition may be enhanced by left ventricular dysfunction of whatever cause in older patients with atrial septal defect.

Whatever the cause of the real and spurious mitral cusp prolapse, this is not related to whether the catheter is passed into the left ventricle across the mitral valve or retrogradely. Experience in assessing mitral anatomy in patients with atrioventricular defects has shown that the catheter route is immaterial; the venous route is preferred where possible, to avoid unnecessary arteriotomy or arterial puncture, particularly in a child.

It is difficult to know why this angiocardigraphic appearance suggesting prolapse is so common in secundum atrial septal defect and yet cannot be confirmed at operation in most patients. So frequently is this radiological appearance seen, that the left ventriculogram which shows it can be considered to be typical of secundum atrial septal defect. When other mitral valve abnormalities were found at operation, these could not be reliably identified on the left ventriculogram. By contrast, when the mitral valve was normal on the left ventriculogram, this observation was reliable and correlated with operative and postoperative findings. We are, therefore, forced to conclude that the radiological appearances on the left ventriculogram suggesting ballooning or prolapse of the posterior mitral cusp, and occasionally the anterior mitral cusp, are common in simple secundum atrial septal defect,

but rarely represent a real pathological abnormality of the mitral valve, except when supporting clinical signs are present.

We are indebted to Dr. Keith Jefferson who criticised us freely, encouraged our work, and allowed us to quote his radiological reports.

We thank Mr. Donald Ross and Mr. John Parker who specifically undertook to inspect and test the mitral valve in each patient at the time of closure of the atrial septal defect

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